CLINICOPATHOLOGICAL STUDY OF DISCOID LUPUS ERYTHEMATOSUS

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CERTIFICATE

This is to certify that this dissertation entitled **" CLINICOPATHOLOGICAL STUDY OF DISCOID LUPUS ERYTHEMATOSUS "** is a bonafide work done by **DR.T.ARUN PRAKASH,** Post Graduate in M.D. Dermatology, Venereology and Leprosy, Madras Medical College, Chennai- 600 003, during the academic year 2006-2008. This work has not been formed previously the basis for the award of any degree.

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DECLARATION

I, **DR.T.ARUN PRAKASH**, solemnly declare that this dissertation titled **"CLINICOPATHOLOGICAL STUDY OF DISCOID LUPUS ERYTHEMATOSUS"** is a bonafide work done by me at Madras Medical College during 2006-2008 under the guidance and supervision of Prof. Dr. B.Parveen, M.D., D.D., Professor and Head, Department of Dermatology, Madras Medical College, Chennai-600 003.

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CONTENTS

INTRODUCTION

Discoid lupus erythematosus is a benign disorder of the skin characterised by well defined, erythematous atrophic plaques covered by a prominent, adherent scale that extends into the orifices of dilated hair follicles. The disease affects twice as many females as males.

The salient histopathological features of classic DLE lesion are hyperkeratosis with follicular plugging, liquefaction degeneration of basal cell layer of epidermis, degenerative changes in the connective tissue and patchy dermal lymphocytic infiltrate around the appendages. Immunohistology shows presence of immunoglobulins IgG, IgA, IgM and complement at the dermo-epidermal junction in the skin lesions.

The majority of researchers consider DLE to be part of a spectrum of the lupus erythematosus diseases (LE). Accordingly, the clinical expression of LE varies from DLE, a benign and strictly cutaneous form, to a systemic form with an unfavorable prognosis, known as Systemic Lupus Erythematosus (SLE). The risk of developing overt SLE is only approximately 6.5%. The risk is higher in patients with Disseminated DLE (22%) than in DLE confined to the head and neck (1.2%) .

REVIEW OF LITERATURE

Hippocrates, Rogerius, Paracelsus, Manardi, Amatus Lusitanus, Sennert are some of the early renowned physicians who have described "Lupus" which is derived from the Latin word which means "Wolf" [1] depicting its nature of destruction.

Lupus Erythematosus (LE) was identified only as a cutaneous disease until a century ago. At the beginning of the last century, blood vessels and connective tissues distributed throughout the body came to be implicated in the pathogenesis which led to the concept of "multisystem malady"[2] .In 1942, Klemperer et al were struck by many morphological features that were common to diseases such as lupus erythematosus, scleroderma, dermatomyositis, rheumatoid arthritis, acute rheumatic fever and polyarthritis nodosa and classified them as "collagen disease" or "collagenosis" [1] .

HISTORICAL BACKGROUND:

In 1826 Rayer[1] described Lupus Erythematosus as "Flexus Sebaceui". In 1828 Biett[3] described the condition as "Erythema Centrifuge". In 1845 Hebra[1] described it as "Seborrhea Congestiva" in which the simile "Butterfly Rash" was used for the first time. In 1851, Cazenave first used the term "lupus erythemateaux" meaning "red wolf" in order to distinguish this disease from

lupus vulgaris[4].In 1875, Kaposi differentiated LE into two forms namely the discoid form and lupus erythematosus disseminatus[3].

In 1963, Edmund Dubois was among the first to use the "Spectrum" analogy for LE, emphasizing that this illness represented a disease continuum extending from the localized DLE at the more benign pole to fully expressed SLE at the more severely affected pole[5]. Dubois' spectrum concept influenced Jim Gilliam's thinking concerning the classification of Cutaneous LE.

DEFINITION:

Discoid Lupus Erythematosus is a benign disorder of the skin characterized by well demarcated, erythematous, slightly infiltrated, "discoid " plaques that often shows adherent thick scales that extend into orifices of dilated hair follicle. The lesion heals with atrophy, scarring and pigmentary changes. The term "DLE" has been used synonymously with the term Cutaneous Lupus Erythematosus.

N.Gilliam divides the cutaneous manifestation of LE into those that shows characteristic histologic changes of LE (LE-Specific skin disease) and those that are not histopathologically distinct for LE and / or may be seen as feature of another disease process (LE-Non specific skin disease).

The term Cutaneous LE is often used synonymously with LE-specific skin disease as an umbrella designation for the three major categories of LE-

specific skin disease: Acute Cutaneous Lupus Erythematosus (ACLE), Subacute Cutaneous Lupus Erythematosus (SCLE), and Chronic Cutaneous Lupus Erythematosus (CCLE). DLE lesions are the most common form of CCLE.

TYPES OF DLE:

Discoid Lupus Erythematosus is classified clinically into the following types[6] .

1.Classic Discoid LE (DLE)

a. Localised DLE

b. Generalised DLE

2.Hypertrophic / Verrucous DLE

3.Lupus profundus / Lupus panniculitis

4.Mucosal DLE

a.Oral DLE

b.Conjunctival DLE

5.Lupus tumidus

6.Chilblain LE

7.Lichenoid DLE (LE / Lichen planus overlap)

EPIDEMIOLOGY OF DLE:

DLE generally affects twice as many females as males, with a peak age of onset in the fourth decades in female and slightly later in males[7]. It is only half as frequent in black people[8]. In a series of 1045 cases, 3% began under 15 years of age and 2.5% at over 70 years[9]. A family history was found in 4% of patients in one series[10].

AETIOLOGY AND PATHOGENESIS:

The causes and pathogenic mechanism responsible are not fully understood. An attractive contemporary model describes four theoretical temporally sequential phases that are prerequisite for the clinical expression of this disease. These phases are inheritance of susceptibility genes, induction of autoimmunity, expansion of autoimmune processes and immunological injury[11].

Genetic susceptibility is probably related to genes that decrease the threshold for self-reactivity and allow a sustained immune response after the first response or the type and extent of end-organ damage[12]. The autoimmune induction phase refers to the development of self-reactive T cells that demonstrate loss of self-tolerance[13]. Mechanisms of induction may include molecular mimicry induced by infectious antigens, abnormal expression of human leucocyte antigen DR, malfunction of thymic or peripheral tolerance,

and presentation of cryptic peptides during apoptosis. The expansion phase involves the progressive development of abnormal autoimmune clonal expansion of B and T cells that have self-recognition capabilities. The final injury phase results in clinical disease and is likely related to autoantibodies and immune complexes causing tissue damage. Mechanisms likely include direct cell death, activation of the complement system, opsonisation and the inhibiting function of target molecule[13].

Genetic Factors:

Genetic factors, including somatic mutation are implicated in the pathogenesis of the disease. Positive associations with HLA-B7, -B8, -Cw7, -DR2, -DR3 and DQW1 are reported[14]. The relative risk is increased with certain combinations of antigens –HLA-CW7, -DR3 and -DQW1 and for HLA-B7, -CW7 and –DR3. The extended haplotype –HLA*01, B*08, DRB1*0301- is associated with both SCLE and DLE, and the A*03, B*07, DRB1*15 haplotype has been associated with DLE lesions[14] .

Environmental Factors:

Genetic predisposition for a lupus diasthesis does not, in itself, produce disease. Rather , it appears that induction of autoimmunity in such patients is triggered by some inciting event, likely an environmental exposure.

The onset of lesions may be precipitated by a variety of factors. These include trauma[15] (11%), mental stress (12%), sunburn (5%), infection (3%), exposure to cold (2%), pregnancy (1%), exposure to x-rays, diathermy, chemical burns, PUVA therapy, laser therapy and drugs[16] (e.g. Isoniazid, Penicillamine, Griseofulvin, Dapsone).

Patients with the primary antiphospholipid antibody syndrome may also develop DLE[17]. The findings of antibodies to reovirus RNA in 42% of patients suggest that viruses may have a role in DLE[18].

HISTOPATHOLOGY:

In most instances of discoid lesions, diagnosis of lupus erythematosus is possible on the basis of a combination of histologic findings. The various clinical types of LE shows an essentially similar histological picture.

Changes may be apparent at all levels of the skin, but all need not be present in every case. The findings are summarized below.

- Stratum corneum: Hyperkeratosis with follicular plugging.

- Thinning and flattening of the stratum malpighii, hydropic degeneration of basal cells, dyskeratosis and squamatisation of basilar keratinocytes.
- Basement membrane: thickening and tortuosity
- Stroma: a predominantly lymphocytic infiltrate arranged along the dermal – epidermal junction, around hair follicle and other appendages and in an interstitial pattern; Interstitial mucin deposition; edema, vasodilatation, and slight extravasations of erythrocytes.
- Pilosebaceous atrophy is a characteristic feature.
- Subcutaneous: Slight extension of inflammatory infiltrate may be present.

The presence of at least two of the following three features is essential to the histological diagnosis of LE[19].

1. Liquefaction degeneration of the basal layer of the epidermis.
2. Degenerative changes in the connective tissue, consisting of hyalinization, oedema and fibrinoid change, most marked immediately below the epidermis.
3. A patchy dermal lymphocytic infiltrate with a few plasma cells and histiocytes, particularly around the appendages, which may be atrophic

Hypertrophic / Verrucous Lupus Erythematosus

Histologically, the epidermis shows hyperkeratosis and papillomatosis. Large number of dyskeratotic keratinocytes are usually noted in the lower portion of the epidermis associated with a band-like mononuclear infiltrate along the dermoepidermal junction. A second pattern consists of a cup-shaped keratin –filled crater surrounded by an acanthotic epidermis with elongated rete ridges and a sparse mononuclear infiltrate[20].

Tumid Lupus Erythematosus:

Histologically, superficial and deep dermal perivascular, interstitial and periappendageal lymphocytic infiltrates associated with stromal mucin deposits are observed[21].

Overlap syndrome- Lichen Planus / Lupus erythematosus :

In some cases of the overlap syndrome, the histologic features and direct immunofluorescence findings are more consistent with lichen planus. In others, the immunofluorescence testings favour lupus erythematosus[22], and still in another subset of patients there are lesions of lichen planus that coexist, rather than overlap, with those of lupus erythematosus[23,24] .

Lupus Erythematosus Profundus/Panniculitis:

Salient histologic findings include a predominantly lobular lymphohistiocytic infiltrate often with plasma cells occasionally forming

germinal centers. Vascular changes include endothelial prominence, thrombosis, calcification or perivascular fibrosis ("Onion Skin" appearance).

IMMUNOPATHOLOGY:

Direct immunofluorescence (DIF) testing shows the presence of immunoglobulins IgG, IgA, IgM and complement at the dermoepidermal junction in skin lesions present for 6 weeks or more, in approximately 80% of patients [25]. They are more frequent on the face and in untreated lesions, and decrease after treatment with topical corticosteroid. Homogenous, granular or thread patterns occurs, but the deposition is usually homogenous in older lesions. Uninvolved skin is always negative, and the biopsy should always be from lesional skin of a well established inflammatory lesion.

CLINICAL FEATURES:

Lesions begin as dull red macules, papules or well defined plaques that develop an adherent scale and evolve with atrophy, scarring, and pigmentary changes. In darker skinned individuals, lesions technically demonstrate areas of both hyper- and de-pigmentation. In lighter skinned patients, the plaques may appear grey or have little pigment alteration.

There is adherent scale in many cases, and when this is removed its undersurface shows horny plugs, which have occupied dilated pilosebaceous canals. This is called 'tin-tack' sign or carpet tacks sign or langue au chat (cat's tongue) sign[26]. In a study at Leeds, 14% had Raynaud's phenomenon and 22% had Chilblains. Joint pains are complained of by approximately 25% of patients.

Most patients have no symptoms of systemic upset, even with widespread cutaneous disease. In patients with Localized Discoid Lupus Erythematosus, the disease is limited to the head and neck.

In patients with Disseminated Discoid Lupus Erythematosus, discoid lesions are seen predominantly on the upper trunk and upper limbs, usually but not always in association with lesions on the head[27].

Localized Discoid Lupus Erythematosus:

Discoid lesions are usually localised above the neck. Favoured sites are the scalp, bridge of the nose, malar areas, lower lip, and ears. The concha of the ear and external canal are frequently involved. Some patients present with periorbital edema and erythema.

On the scalp, most lesions begin as erythematous patches or plaques that evolve into white often depressed, hairless patches. Alopecia occurs in the scalp lesion in approximately one-third of patients[28].

There are several clinical variants. If hyperkeratosis is marked, warty lesions with a red, slightly raised edge results. This warty type of LE is most commonly seen on the nose, temples, ears, and scalp, but may also occur on the palms and soles[29].

Non-pruritic papulonodular lesions may occur on the arms and hands, resembling keratocanthoma or hypertrophic Lichen planus[30]. Histological examination of these lesions typically demonstrate a lichenoid dermatitis. The lips and scalp may also demonstrate lesions that resemble lichen planus or lichen planopilaris. This variant is called Verrucous Lupus Erythematosus[26].

Sometimes, the appearance may be psoriasiform[31]. Lesions may be present as reddish, well defined almost smooth plaques with little or no scaling[31]. Sometimes these plaques may show prominent flattening in the center, giving rise to annular lesions.

A pruritic chronic umbilicated papular eruption may occur on the back and results in acneiform hypertrophic follicular scar[32].

In 7.5% of patients, the lesions on the face resemble rosacea. LE of this type presents with reddish nodular lesions on the nose, cheeks, forehead, and sometimes chin, and is associated with a diffuse erythema of the face and easy flushing. Usually, there are no pustules as in true rosacea[35].

Another clinical type of DLE results in annular atrophic plaques on the face, neck, and behind the ears[36]. The centers of the plaques is depressed and sclerotic, and the lesions resemble morphea, lichen sclerosus or annular atrophic plaques[37].

Disseminated DLE:

Disseminated DLE is less common than localized DLE[38]. All degrees of severity are encountered. Most often the thorax and upper extremities are affected in addition to the head and neck. This occurs almost always in women and they are usually smokers. This variety tends to be persistant, resistant to therapy and associated with severe psychological upset[39].

Laboratory abnormalities such as an elevated erythrocyte sedimentation rate, elevated antinuclear antibodies, single stranded (ss) DNA antibodies and leucopenia are more common with this form of LE than with localized DLE[38]. The clinical presentation of Disseminated Discoid Lupus Erythematosus may vary.

In 'Lupus Erythematosus Telangiectoides' reticulate telangiectasia are seen on the arms, legs, and back of the calves[40]. This clinical variant occurs in SLE as well as DLE[41]. The appearance are characterized by a persistent blotchy reticulate telangiectasia, which occurs on the face, neck, ears, dorsa of the

hands, breast, heels, and on the sides of the feet[41]. Healing occurs with punctate atrophic scarring.

A more annular variant has been called 'Lupus Erythematosus Gyratus Repens' and consists of a migratory gyrate annular erythema with the histological features of LE, although lupus band test is negative[42]. There may be an underlying carcinoma.

Rarely bullous lesions may occur[43]. Occasionally, one or more fingers may show a curious atrophic spindling, sometimes with hyperextension of the terminal phalanges and dystrophy of nails[44].

Arteritic lesions resembling those of Dego's syndrome or disseminated atrophic blanche occasionally occur, and linear lesions following Blaschko's lines have been reported[45].

Chilblain Lupus:

Some LE patients develop red-purple patches and plaques on their toes, fingers, and face that are precipitated by cold, damp climates. Such lesions are highly reminiscent of simple chilblains or pernio lesions[47]. As these lesions evolve, however, they take on the typical appearance of DLE lesions clinically and histopathologically[46]. It can be precipitated by pregnancy[48]. Chilblains lupus patients often have typical DLE lesions on the face and head[47]. Less commonly, discoid and perniotic lesions start simultaneously, and sometimes

perniotic lesions occur alone. A study found Ro / ss-A antibodies in the sera of eight out of nine Chilblain lupus patients, suggesting that these antibodies might be a useful clinical marker of this disorder[49]. They are also either smokers, or have markedly abnormal peripheral circulation with low resting volume[50].

Tumid LE

The dermal form of LE without surface/epithelial changes is known as Tumid LE. Clinically affected patients display indurated papules, plaques, and nodules without erythema, atrophy or ulceration of the surface[21]. Dermal mucin deposition is typical. The lesions generally respond readily to antimalarials[26].

Mucosal DLE

Burge and coworkers have confirmed that the prevalence of mucous membrane involvement in Chronic Cutaneous LE is about 25%[51]. While oral mucosa is most frequently involved, nasal, conjunctival and genital mucosal surfaces also can be affected[51].

Within the mouth, the buccal mucosal surfaces are most commonly involved, with the palate, alveolar process and tongue being sites of less frequent involvement[51]. Individual lesions begin as painless, erythematous patches later maturing to a chronic plaque that can present an appearance quite similar to that of lichen planus. The chronic buccal-mucosal plaques have a sharply marginated, irregular scalloped white border with radiating white striae

and telangiectasia[51]. The surface of these plaques overlying the palatal mucosa often has a well-defined meshwork of raised hyperkeratotic white strands that encircle zones of punctate erythema, which gives a "honey comb" appearance. The centre of older lesions can become depressed and occasionally undergo painful ulceration.

Well-defined chronic DLE plaques also can appear on the vermillion border of the lips. At times, DLE involvement of the lips can present as a diffuse chelitis, especially on the more sun exposed lower lip. Although lesions can appear on the tongue, this location is quite rare[51]. Oral lesions may resemble leukoplakia[52]. Erythematous lesions occur on the vulva in 5%[42] or around the anus[53].

Eye Lesions:

Conjunctival DLE lesions begins as small areas of inflammation most commonly affecting the palpebral conjunctivae or the margin of the eyelid. The lower lid is affected more often than the upper eyelid. Palpebral lesions have been reported without lesions elsewhere on the face[54]. The eyelids are red, especially peripherally and are slightly infiltrated and always scaly[54]. The lesions may itch, and are exacerbated by trauma and sunlight. Corneal involvement is rare. Superficial punctate keratopathy and stromal keratitis have been reported[55]. Acute mucinosis of the eyelids and periorbital skin can occur[56].

Lupus Erythematosus and Erythema Multiforme-Like Syndrome (Syn. Rowell's Syndrome):

The distinct syndrome of cutaneous LE, either discoid or systemic, occurring with lesions resembling erythema multiforme on the face, neck, hands, chest, and in the mouth was first described by Rowell in 1963 in patients with discoid LE, but may be seen in both subacute and systemic disease[57]. Characteristically, it lasts from a few days to over a month, but episodes may occur at intervals over a period of 20 years[57]. The lesions are at first papular, but later a ring forms and the edge becomes vesicular. Bullae, necrosis, and ulceration may develop if the reaction is intense, although sometimes healing occurs without scarring. Patients with this syndrome also frequently have perniotic lesions[58].

These patients show a characteristic pattern of serological abnormality, in that the speckled type of antinuclear factor is associated with Rheumatoid factor and the precipitating antibody to saline extract of human tissues (anti-SjT)[59]. When the syndrome occurs in DLE, the dermal-epidermal band test is positive in the discoid lesions and negative in the erythema multiforme lesions[60].

Childhood DLE:

DLE is uncommon in childhood[62]. There appears to be no female preponderance, there is less photosensitivity and the frequency of progression to systemic disease is higher[62]. The other clinical features are similar to those of adults.

Lichen planus – Lupus erythematosus Overlap Syndrome :

This variant is characterised by lesions that share features of lichen planus and lupus erythematosus. Atrophic plaques and patches with hypopigmentation and a livid red to blue-violet colour with telangiectasia and minimal scaling are characteristic. Transient bulla may develop. Classic lesions of lichen planus are not usually seen. Photosensitivity, pruritus and follicular plugging are also not common. Lesions may develop anywhere, but are most common on the extremities. Some patients with this overlap syndrome may progress to systemic lupus erythematosus. In other instances, laboratory evaluation may reveal only a weak-positive antinuclear antibody. This disease variant is characterised by a prolonged course and lack of response to treatment.

In some cases of the overlap syndrome, the histologic features and direct immunofluorescence findings are more consistent with lichen planus. In others, the immunofluorescence testings favour lupus erythematosus[22], and still in another subset of patients there are lesions of lichen planus that coexist, rather

than overlap, with those of lupus erythematosus[23,24] . By direct immunofluorescence, the most common finding is the presence of cytoids staining with IgG, IgM and C3 intraepidermally or at the dermal-epidermal junction, as seen in classic lichen planus. Linear to granular deposition of IgM and C3 (as seen in lupus erythematosus, but not in lichen planus) has been observed occasionally. Shaggy deposition of fibrinogen at the basement membrane zone, typical of lichen planus, is sometimes found[61].

Lupus Erythematosus Profundus / Panniculitis:

Historically, referred to as Kaposi-Irgang disease[63], this rare form of Chronic Cutaneous LE is characterized by inflammatory lesions in the lower dermis and subcutaneous tissue. Approximately 70% of the patients with this type of Chronic Cutaneous LE also have typical DLE lesions often overlying the panniculitis lesions[64]. The term "LE profundus" has been used by some to arbitrarily designate those patients who have both LE panniculitis and DLE lesions.

The lesions present as deep, firm, 1-3 cm diameter nodules often with normal appearing overlying skin[65]. The skin ultimately becomes attached to the firm, subcutaneous nodular lesions and is drawn inward to produce deep, saucerized depressions as the lesions mature[65]. The head, proximal upper arms, chest, buttocks and thighs are the sites of predominant involvement. Confluent

involvement of the face can stimulate the appearance of lipoatrophy. LE profundus can also present as periorbital edema[66,67].

Linear involvement of the extremities also has been observed[68]. Dystrophic calcification within older lesions of LE profundus is common and at times can be a prominent clinical feature of the disease requiring surgical excision. In addition, LE panniculitis may produce breast nodules that can mimic carcinoma clinically and radiologically[69].

Associated Features:

Small telangiectases on the face occur in approximately 20% of the patients[70]. These dilated vessels are usually small capillaries of irregular size and distribution, particularly on the cheeks. Dilated nail fold capillaries similar to those seen in SLE and dermatomyositis, may be visible with naked eye[70]. More than half of the patient states that they have dry skin. Occasionally mild diffuse alopecia may occur. Alopecia areata has occurred in 10% of one series of DLE[71].

Bilateral enlargement of the parotids has been reported[72, 73]. Histologically there is lymphocytic infiltration of the parotid, which is said to be like that of LE profundus[73] . Livedo reticularis on the legs has been reported in DLE[74].

Porphyria cutanea tarda and, less commonly, variegate porphyria are acute intermittent porphyria may occur in patients with DLE[75, 76]. The relation to pemphigus, myasthenia gravis and thymoma has been discussed[77].

DLE has also been associated with chronic lymphatic leukemia, macroglobulinaemia[78], polychondritis[79], autoimmune thyroiditis[80], carpal tunnel syndrome[81], PLE[82], Sheehan's syndrome[83] and erysipelas[84]. Hereditary C2 deficiency occurs in association with skin lesions resembling the discoid lesions of SLE[85]. Hereditary deficiency of the third and of the fifth component of complement has also been associated with a lupus like syndrome[86]. DLE has also occurred with C1q deficiency[87].

LABORATORY ABNORMALITIES IN DLE:

Abnormalities were found in 55% of patients[88]. Anaemia, leucopenia, or thrombocytopenia can be found in approximately one third of the patients and erythrocyte sedimentation rate is raised in 20%[88]. The serum globulin is raised in 29% of the patients[88], elevation of gammaglobulin being the most common abnormality[88]. Higher IgG levels are associated with scarring[89]. Occasionally the Coomb's test may be positive and cryoglobulins [90, 91] and cold agglutinins may be detected in the serum.

False positive reactions for syphilis have been reported in 26% of cases[92]. Anticardiolipin antibodies (mainly IgM) in low titers occur in approximately 15% of patients[93]. Sometimes LE cell test may be positive (1.7%). Rheumatoid factor is present in approximately 17% of patients[88].

Antinuclear antibodies are found in 35%, the 'homogenous' type of antinuclear factor being twice as frequent as the 'speckled' type[89]

Antinuclear antibodies are more common in older patients, in those who have had the disease for a long time and when there is extensive skin involvement. They are also more common in patients with Chilblains, Raynaud's phenomenon and joint pains[90]. The incidents of anti-DNA antibodies varies from 0% to 27%[91-93] .

Antibodies to RNA occur in 42%[94]. Lower titer anti-RO antibodies are found in 10% of the patients with DLE[95]. Serum complement levels are occasionally reduced[91]. Antibodies to the saline extracts of liver, blood vessels, skin, kidney, heart, spleen, leukocytes, gammaglobulin and deoxyribonucleoproteins have been detected by complement utilization[96]. The highest titres occur in the mildest non-scarring forms of the disease and lowest in the scarring type. It has been suggested that these antibodies may have a protective function[96].

A high incidence of antithyroid antibodies has been found in DLE, particularly in females[97]. Gastric parietal cell cytoplasmic antibodies occur in 13% of the patients[97].

T-cell counts are significantly lower than in controls, although B-cells are not reduced[98].

DIFFERENTIAL DIAGNOSIS:

With respect to diagnosis of classical Discoid Lupus Erythematosus, discoid-shaped skin lesions that have erythema and hyperpigmentation at their active borders and depigmentation, telangiectasia and atrophy at the centers are very unlikely to result from dermatological disorders other than cutaneous LE.

Polymorphous light eruption can be expressed in several clinical forms including succulent red plaques that occasionally can mimic earlier phases of evolving DLE lesions[99]. PLE and DLE may coexist, or PLE may precede DLE by many years[99] .

Sarcoidosis, Jessner's benign-lymphocytic infiltration of the skin, pseudolymphoma of Spiegler-Fendt, lymphocytoma cutis, angiolymphoid hyperplasia with eosinophilia, lymphoma cutis, lupus vulgaris[100] and tertiary syphilis[101] are other disorders that can clinically simulate some phases of DLE lesions and at times present diagnostic confusion.

Necrobiosis lipoidica can give rise to facial lesions like DLE. The rosaceous type of LE can usually be differentiated from true rosacea by the absence of pustules. Granuloma faciale also can present as indolent facial plaques.

Hypertrophic Discoid Lupus Erythematosus could be mistaken for keratoacanthoma, squamous cell carcinoma, prurigo nodularis or hypertrophic lichen planus[102].

The differentiated diagnosis of patients with lupus panniculitis includes Weber-Christian panniculitis, factitial panniculitis, pancreatic panniculitis, traumatic panniculitis, morphea profundus, eosinophilic fascitis, sarcoidosis, subcutaneous granuloma annulare and rheumatoid nodules[103]. Oral lichen planus presents the closest clinical appearance to that of oral mucosal DLE[104].

PROGNOSIS:

Lesions of long standing with much scaling and some scarring are slower to remit[105]. Ultimately scarring is found in 57% with scarring alopecia in 35% and pigmentary abnormalities in 35% [106]. Complete remission in the course of years can be expected in over 50%[105].

Long duration and lack of remission are related to Raynaud's phenomenon, scalp involvement and Chilblain-like lesions[107]. Relapses occurring with sunlight, cold, trauma or mental stress after months or years of

remission are not frequent. Inspite of the chronic and relapsing nature of the condition, the patient usually remains in good health.

The risk of developing overt SLE is approximately 6.5%[107, 108]. The risk is higher in patients with disseminated DLE (22%) than in DLE confined to head and neck (1.2%).

Squamous cell and less commonly basal cell carcinomas occasionally occur in scars of DLE. An incidence of 3.3% has been noted[109].

TREATMENT:

General measures play a large part in successful management. Patient should be warned against excessive exposure to sunlight. A sunscreen cream or lotion should be prescribed[110]. A preparation with a UVB protection factor of atleast 15 is required, but UVA protection is also important. Application should be frequent- probably every 2-3 hours in summer.

Initial treatment usually includes the use of a potent topical corticosteroid like clobetasol propionate 0.05%, betamethasone dipropionate 0.05% or diflorosone diacetate 0.05%[111].

Intralesional corticosteroid injections are helpful in resistant cases[112,113]. Intralesional injections of antimalarials including chloroquine[114], have been tried but results were not as good as with corticosteroids. Interferon-α (IFN-α) has also been used intralesionally with success[115]. Among other local measures,

cryotherapy, surgical excision[116], painting small lesions with trichloroacetic acid and local laser therapy may be helpful. The carbondioxide laser, pulsed-dye and argon lasers may be valuable for telangiectatic LE[117]. For patients with severe, extensive or scarring disease, particularly affecting the scalp, oral prednisolone at 0.5 mg/kg rapidly tapered over 6 weeks is quickly effective.

One or combination of the aminoquinoline antimalarials can be effective for approximately 75% of patients with cutaneous LE. Hydroxychloroquine, chloroquine sulphates[118], mepacrine[118] has helped DLE lesions. Cigarette smoking reduces the efficacy of treatment with antimalarials, probably by modifying metabolism[119].Monitoring during therapy should include taking an ophthalmic history, and testing reading ability with appropriate charts[118].

Other systemic drugs found useful include Acitretin, Etretinate, Isotretinoin, Dapsone, Methotrexate, Auronafin, Thalidomide, Clofazimine, Danazol, Sulfasalazine, Phenytoin, Beta-carotene, Cyclophosphomide, Azathioprine, Gold by intramuscular injection[120-138]. Excision without grafting was successful in a case of verrucous LE following burn[139].

AIM OF THE STUDY

1. To study the incidence of Discoid Lupus Erythematosus in Government General Hospital, Chennai during the period between July 2006 to September 2008.
2. To study the incidence of various clinical types of Discoid Lupus Erythematosus.
3. To study the age and sex distribution.
4. To study the commonest site of lesions.
5. To study the main presenting complaints and precipitating factors.
6. To study the relevant serological abnormalities.
7. To study the associated disorders.
8. To study Histopathological features in all cases and immunofluorescence pattern in selective cases.

MATERIALS AND METHODS

All the patients attending the outpatient department of Government General Hospital, Chennai during the period between July 2006 to September 2008 were screened and patients with morphology suggestive of Discoid Lupus Erythematosus were enrolled in the study. There were **51** patients who were clinically diagnosed as Discoid Lupus Erythematosus out of 30,056 patients attending Dermatology OP during the study period. A detailed history as given in the proforma was elicited. Various presenting complaints like photosensitivity, Raynaud's phenomenon, joint pain, loss of hair were also recorded. Biopsy was done on all patients. All patients were subjected to various investigations like Total count, differential count, hemoglobin, erythrocyte sedimentation rate, blood sugar, blood urea, liver function test, Antinuclear antibody titre, Rheumatoid factor, 'C' reactive proteins, and anti-double stranded DNA antibody. Direct Immunofluorescence was done for few patients. Opinions of Rheumatologists were obtained in patients with joint pain.

OBSERVATIONS AND RESULTS

Incidence:

Of the total 30,056 patients attending Skin OP, Government General Hospital, Chennai during the period between July 2006 to September 2008, total number of patients with Discoid Lupus Erythematosus was 51.

Incidence of Discoid Lupus Erythematosus was 1.7 per 1000 that accounts to 0.17%.

TABLE-1

Total number of patients attending Skin OP in Govt. Gen. Hospital (July 2006 to September 2008)	30,056
Total number of patients with Discoid Lupus Erythematosus	51
Incidence of Discoid Lupus Erythematosus	0.17%

AGE AND SEX WISE DISTRIBUTION

Youngest age at presentation was in a 16 year old female. Oldest age at presentation was in a 65 year old woman. Incidence of DLE peaked between 31-50 years. DLE was more common in females (69%) than males (31%). The sex ratio between female and male was 2.3 : 1 in this study.

TABLE-2

Age in years	Male	Female	Total
0-10	0	0	0
11-20	0	3	3
21-30	2	6	8
31-40	4	11	15
41-50	6	9	15
51-60	4	4	8
61-70	0	2	2
Total	16	35	51

AGE AND SEX WISE DISTRIBUTION OF LOCALIZED DLE

Peak incidence was between 31-40 years. Peak incidence among males was between 41-60 years & among females was between 31-50 years. Localized DLE was more common in females (77%) than in males (23%). The sex ratio between female and male was 3.5 : 1 in this study.

TABLE-3

Age in years	Male	Female	Total
0-10	0	0	0
11-20	0	3	3
21-30	1	6	7
31-40	2	10	12
41-50	3	8	11
51-60	3	3	6
61-70	0	0	0
Total	9	30	39

AGE AND SEX WISE DISTRIBUTION OF DISSEMINATED DLE

Incidence of Disseminated DLE peaked between 41-50 years. Incidence of Disseminated DLE in males (58%) was more common than in females (42%). The sex ratio between female and male was 1 : 1.3 in this study.

TABLE-4

Age in years	Male	Female	Total

0-10	0	0	0
11-20	0	0	0
21-30	1	0	1
31-40	2	1	3
41-50	3	1	4
51-60	1	1	2
61-70	0	2	2
Total	7	5	12

CLINICAL TYPES OF DLE AND THEIR INCIDENCE

Total number of Cases : 51

TABLE-5

S.No	Clinical Types	No. of Cases	Percentage
1	Localised DLE (Head & Neck)	39	76%
2	Disseminated DLE (Trunk & Extremities in	12	24%

	addition to Head & Neck)		

Localized DLE was 3 times more common than disseminated DLE.

LOCALIZED DLE

Localized DLE constituted 76% of the total number of cases. Among Localized DLE, the incidence of various clinical presentations are as follows:

TABLE-6

S. No	Clinical Presentation	No. of Cases	Percentage
1	Localized Discoid lesions	22	56%
2	Localized Discoid lesions + Mucosal lesions	6	15%
3	Localized Discoid lesions + Follicular pits in ears	7	18%

4	Hypertrophic / Verrucous DLE	1	3%
5	Mucosal lesions alone	2	5%
6	Eye lesions	1	3%
Total		39	76%

The commonest clinical presentation of Localized DLE was Localized circumscribed or discoid type of lesions (56%).

DISSEMINATED DLE

Disseminated DLE constituted 24% of the total number of cases. The incidence of various clinical presentations are as follows:

TABLE-7

S. No	Clinical Presentation	No. of Cases	Percentage
1	Disseminated Discoid lesions	6	49%
2	Disseminated Discoid lesions + Mucosal lesion	4	33%
3	Lupus Erythematosus - Lichen Planus overlap syndrome	1	9%

4	LE gyratus repens	1	9%
Total		12	24%

MUCOSAL DLE - INCIDENCE

Incidence of Mucosal lesions in DLE : 27%.

Incidence of involvement of only the Mucosa : 4%.

Incidence of Mucosal lesions with cutaneous discoid lesions : 23%.

Mucosal involvement in females (28%) were more common than in males (25%). The sex ratio between female and male was 1.1 : 1 .

Patient with Disseminated DLE had higher incidence (42%) of mucosal lesions than those with localized DLE (18%).

TABLE- 8

Clinical Presentation	Total No. of Cases	Total

	Male	Female	
Mucosal lesions alone	0	2	2
Mucosal lesions along with Localized DLE	1	6	7
Mucosal lesions along with Disseminated DLE	3	2	5
Total	4	10	14

TYPES OF MUCOSAL LESIONS

Involvement of the lower lip was the commonest (16%) of the mucosal lesion.

TABLE-9

S. No	Mucosal Lesion	No. of Cases
1	Erosions and crusting of Lip	8
2	Hyperpigmented patches on the Buccal Mucosa	3
3	Erosions on the Palate	2
4	Erosions over the Lips and Palate	1
Total		14

PRESENTING COMPLAINTS

The most common presenting complaint was Burning Sensation (39%) on exposure to sunlight.

TABLE-10

S. No	Presenting Complaint	No. of Patients
1	Itching	15
2	Burning Sensation	20
3	Loss of hair	5
4	Cosmetic disfigurement	5
5	Scaling	3
6	Pain	3

PRECIPITATING OR EXACERBATING FACTORS

The most common exacerbating factor was sunlight (57%).

TABLE-11

S. No	Precipitating or Exacerbating Factor	No. of Cases
1	Sunlight	29
2	Trauma	1
3	Mental Stress	2
4	Burns	1
5	Pregnancy	1
6	Drugs	2

7	Premenstrual Flare	1

OCCUPATION OF THE PATIENT

31% of the patients were agriculture labourers.

10% of the patients were watchmen.

TABLE-12

OCCUPATION	MALE	FEMALE	PERCENTAGE
Agriculture	4	12	31
Housewife		20	39
Watchman	5		10
Shopkeeper	2		4
Student		1	2
Teacher	1	1	2
Driver			2
Tailor	1	1	2
Mason	1		2
Office clerk	1		2
Plumber	1		2

Photographer	1		2
Total	16	35	

SITE OF INVOLVEMENT

LOCALISED DLE

Total number of cases : 39

Most common site involved in Localized DLE - scalp (38%)

TABLE-13

S.No	Site	No. of Cases
1.	Scalp	15
2.	Face	4
3.	Lips	2
4.	Ears	3
5.	Multiple Sites (Scalp, Face, Ears, Lips)	15

DISSEMINATED DLE

Trunk was involved in 92% of Disseminated DLE Cases along with lesions on the head.

Trunk alone was involved in a single case.

TABLE-14

S.No.	Site	No. of cases
1.	Head, Trunk & Upper limbs	2
2.	Head, Trunk ,Upper &	2

	lower limbs	
3.	Head & Trunk	6
4.	Head & Upper limb	1
5.	Trunk alone	1

RAYNAUD'S PHENOMENON:

No. of cases with H/o Raynaud's phenomenon : 3

Percentage of cases with Raynaud's phenomenon : 6%

TABLE-15

Type	Raynaud's phenomenon+	Total no. of cases	%
Localized DLE	1	39	2.5 %
Disseminated DLE	2	12	17 %
Total	3	51	6 %

JOINT PAIN:

12% of Cases (6) had complaints of joint pain. Joint pain was more common in Disseminated DLE than in Localized DLE.

TABLE-16

Type	No. of Cases	Total no. of	%

	with joint pain	cases	
Localized DLE	2	39	5 %
Disseminated DLE	4	12	33 %
Total	6	51	12 %

ASSOCIATED CONDITIONS

TABLE - 17

Associated Conditions	No. of Patients
Polymorphic Light Eruption	1
Diffuse Hair Loss	1
Urticaria	1
Prurigo nodularis	1
Macular Amyloid	1
Hypothyroidism	2
Diabetes Mellitus	2
Hypertension	1
Pterigium of Eye	1

LABORATORY ABNORMALITIES IN DLE

ANAEMIA:

Percentage of patients with anaemia were found to be 25%

TABLE – 18

Type	No. of Patients with anaemia	Total no. of Pts	%
Localized DLE	10	39	26 %
Disseminated DLE	3	12	25%
Total	13	51	25%

ERYTHROCYTE SEDIMENTATION RATE (ESR):

Percentage of patients with raised ESR was 57%

TABLE – 19

ESR >20mm/hr	No. of Patients
Total no. of patients investigated	51
Total no. of patients with raised ESR	29
Percentage of patients with raised ESR	57%

OTHER PARAMETERS:

- Total Leukocyte count, platelet counts were within normal limits in all patients.
- No abnormality was detected in Renal and Liver Function parameters.
- Serum proteins were normal in all patients.

ANTINUCLEAR ANTIBODY POSITIVITY

Percentage of patients with ANA positivity was found to be 29%

TABLE – 20

Types of DLE	ANA Positivity	Total No. of Cases	Percentage
Localized DLE	10	39	26%
Disseminated DLE	5	12	42%
Total	15	51	29%

RHEUMATOID FACTOR

Percentage of Rheumatoid Factor positivity was found to be 33%

TABLE – 21

Type	RF Positive	Total No. of Patients	Percentage
Localized DLE	11	39	28%
Disseminated DLE	6	12	50%
Total	17	51	33%

C-REACTIVE PROTEIN (CRP)

Percentage of CRP Positivity was found to be 33%

TABLE – 22

Type	CRP Positive	Total No. of Patients	Percentage
Localized DLE	9	39	23%
Disseminated DLE	8	12	67%
Total	17	51	33%

ANTI – DOUBLE STRANDED DNA ANTIBODY was found to be negative in all patients.

NEOPLASTIC CHANGES IN DLE

Only one case presented with malignant change.

One case presented with Squamous Cell Carcinoma complicating Lupus Erythematosus – Lichen Planus overlap Syndrome.

Incidence of Malignancy in DLE : 2%

HISTOPATHOLOGY :

Skin biopsy was done in all the cases. Histopathological study showed compatibility with the features of DLE in all cases, except one. One case showed features suggestive of Lichen planus. Biopsy of the verrucous growth from the lesional skin on the right elbow in this case showed features of well differentiated squamous cell carcinoma. Another case showed features suggestive of Verrucous DLE.

DIRECT IMMUNOFLUORESCENCE :

DIF was done in a few cases . The biopsy was taken from the lesional skin, exposed nonlesional skin and unexposed non-lesional skin. Most of the cases showed moderately strong homogenous IgG, IgM, IgA and C3 at the basement membrane zone band in the lesional skin and negative in the uninvolved skin, features suggestive of Discoid Lupus Erythematosus. One case showed Discontinuous ragged fibrinogen band in the basement membrane zone and IgG, IgM and C3 colloid bodies in the lesional skin. These features were suggestive of Lichen planus.

DISCUSSION

In this study, among the total 30,056 patients attending Skin OP, Government General Hospital, Chennai during the period between July 2006 to September 2008, total number of patients with DLE was 51. The incidence of

DLE was found to be 1.7 per 1000 (Tab.1). A study by G. H. Findlay and J. G. H. Lups of 191 cases, the incidence was found to be 0.07 per 1000 [140] .

In this study, the youngest age at presentation was 16 years and the oldest age was 65 years. Incidence of DLE peaked between 31 – 50 years (Tab.5). This was similar to the study by Damm J and Sonnischsen N where the peak age of onset was 4th decade in female and slightly later in males[9] .

The sex ratio between female and male was 2.3 : 1 in this study (Tab.2). In the study by Damm J and Sonnischsen N, the sex ratio was 2 : 1 . The female to male ratio for Localised DLE in this study was 3.5 : 1 (Tab.3) as compared to 3 : 1 in the study conducted by Tebbe R and Orfanos[141]. The female to male ratio for disseminated DLE was 1 : 1.3 in this study (Tab.4) as compared to 13 : 1 in the study conducted by Tebbe R and Orfanos[141] . There was no incidence of familial cases in this study. In the study by G. H. Findlay and J. G. H. Lups one case of familial occurrence was found in a series of 191 cases[140] . Sonnichsen et al in east berlin have had similar experience. They found only three familial cases in a total of 1,100 affected cases[140] .

Percentage of Localised and Disseminated DLE was 76% and 26% respectively in this study (Tab.5) , where as in a study by Tebbe R and Orfanos of 97 patients the percentage of localized and disseminated DLE was 40% and 60% respectively[141] .

In this study, the percentage of cases with mucosal lesions was 27% (Tab.8) similar to the study conducted by Thai Helena and Nelson Guimaraes Proenca[142] .

Mucosal involvement was seen 28% of females compared to 25% in male patients (Tab.8). Patients with Disseminated DLE had higher incidence (42%) of mucosal lesions than those with localized DLE (18%) (Tab-8).

Involvement of the lips was the commonest (16%) of the mucosal lesion (Tab-9) in this study. In a study by Thai Helena and Nelson Guimaraes Proenca , lips were the commonest site (13%) of mucosal lesion[142] .

The most common presenting complaint was burning sensation (39%) on exposure to sunlight, followed by itching (29%) in this study(Tab-10). In a study by Thai Helena and Nelson Guimaraes Proenca , itch and aggravation following sun exposure (59%) was the commonest complaint, followed by pruritus(42%)[142] .

The most common exacerbating factor was sunlight (57%) in this study followed by mental stress (0.3%), drugs (0.3%), trauma (0.1), burns (0.1%), pregnancy (0.1%) and premenstrual flare (0.1%) (Tab- 11). In a study at Leeds, the most common precipitating factor was mental stress (12%), followed by trauma (11%), sunburn (5%), infection (3%), exposure to cold (2%) and pregnancy (1%)[143] . In this study, 31% of the patients were agriculture cooly

and 10% of the patients were watchman by occupation(Tab.12). Both agricultural labourers and watchman have more exposure to sunlight, which is one of the precipitating and exacerbating factor of DLE.

In this study, Scalp was involved in 38% of the localized DLE cases (Tab-13) similar to the study by Thai Helena and Nelson Guimaraes Proenca where scalp was involved in 31.7% of cases[142] .

Trunk was involved in 92% of Disseminated DLE cases. This study also showed that 0.1% of the cases had lesions exclusively in the trunk (Tab-14) whereas in the study by Thai Helena and Nelson Guimaraes Proenca, 0.3% of the cases had lesions exclusively in areas other than cephalic segment[142] . Trunk was involved in 21% of DLE cases in this study (Tab-14) compared to 32% in the study by Thai Helena and Nelson Guimaraes Proenca[142] .

Raynaud's phenomenon was present in 6% of cases in this study (Tab-15) compared to 14% in the study at Leeds of 120 patients[144] .

12% of cases had joint pain in this study (Tab-16) compared to 24% in this study by Thai Helena and Nelson Guimaraes Proenca[142] .

Associated conditions in this study includes polymorphic light eruption, diffuse hair loss, urticaria, prurigo nodularis, macular atrophy, macular amyloid, hypothyroidism, diabetes mellitus, hypertension and pterigium of the eye (Tab-17). Wojnarowska F reported simultaneous occurrence of DLE and

polymorphic light eruption[82] .Mild diffuse hair loss has been reported by Werth VP, White WL and Sanchez MR[71] . Autoimmune thyroiditis has been reported by Van der meer-Rosen CH, Maes EPJ and Faber WR[80] .

20% of patients in this study were known smokers. An association between DLE and smoking has been reported[146] . Disseminated DLE is reported to occur more commonly in women who are cigarette smokers[39]. A higher prevalence was noted in DLE cases (84.2%) than control (33.5%) in a study[146].

Skin biopsy was done in all the cases. All cases showed features suggestive of Discoid Lupus Erythematosus. One case showed features suggestive of Lichen planus. Biopsy of the verrucous growth from the lesional skin on the right elbow in this case showed features of well differentiated squamous cell carcinoma. Another case showed features suggestive of Verrucous DLE.

DIF was done in a few cases. Most of the cases showed moderately strong homogenous IgG, IgM, IgA and C3 at the basement membrane zone band in the lesional skin and negative findings in the uninvolved skin, features suggestive of Discoid Lupus Erythematosus. One case showed Discontinuous ragged fibrinogen band in the basement membrane zone and IgG, IgM and C3 colloid bodies in the lesional skin. These features were suggestive of Lichen planus.

One of the case showed clinical features suggestive of Disseminated Discoid Lupus Erythematosus. But the histological and direct immunofluorescence findings were consistent with Lichen planus. Hence a final diagnosis of Lupus erythematosus-Lichen planus overlap syndrome was made. This case had a verrucous growth developing from the skin lesion on the right elbow which was histologically a well differentiated squamous cell carcinoma.

Laboratory investigations showed anaemia in 25% and raised ESR in 57% of patients (Tab-18,19). A study by Rowell NR found anaemia, leucopenia or thrombocytopenia in approximately one – third of patients and ESR was raised in 20% of patients[88] . Total leucocyte count, platelet counts were within normal limits in all patients in this study. Serum proteins were normal in all the patients in this study.

ANA positivity was 29% in this study (Tab-20), whereas in the study by Rowell NR 35% of patients showed ANA positivity[88] . In this study, percentage of ANA positivity in localized DLE was 26% compared to 42% in disseminated DLE.

Rheumatoid factor was positive in 33% of patients (Tab-21), whereas in the study by Rowell NR, 17% of patients were positive for rheumatoid factor[88] . C reactive protein positivity was found in 33% of patients in this study (Tab-22). Anti-ds-DNA antibody was negative in all patients in this study similar to the study by Kulick KB, Provost TT and Reichlin M[92] . In another study by

Davis P, Atkin B and Hughes GRV anti-DNA antibodies were found in 27% of the cases[93] .

Malignant change was found in only one case (2%) in this study. In a study by Millard LG and Barker DJ, an incidence of 3.3% was noted in a series of 120 white patients[109] .

CONCLUSION

- The incidence of Discoid lupus erythematosus during the period from July 2006 to September 2008 was 1.7 per 1000.
- The incidence of Localised and Disseminated discoid lupus erythematosus were as follows

  Localised DLE - 76%

  Disseminated DLE – 24%
- The most common clinical presentation of localized DLE was localised circumscribed lesions (59%).
- The maximum number of patients were in the age group of 31 – 50 years.
- The female to male sex ratio was 2.3 : 1
- No familial cases were seen.
- Peak incidence of localised DLE was between 31 to 40 years. Peak incidence of disseminated DLE was between 41 to 50 years.
- Localised DLE was more common in females (77%) than males (23%)
- Disseminated DLE was more common in males (58%) than females (42%).

- Mucosal lesions were found in 27% of the patients. Patients with disseminated DLE had higher incidence of mucosal lesions (42%) than those with localised DLE (18%).
- Mucosal involvement was more common in female patients (28%) than in males (25%).
- Involvement of the lower lip was the commonest of the mucosal lesions.
- The most common presenting complaint was burning sensation (39%) on exposure to sunlight. The most common exacerbating factor was sunlight (57%).
- Scalp (38%) was the most common site involved in localized DLE. Trunk was involved in 92% of disseminated DLE patients.
- Raynaud's phenomenon was present in 6% of the patients. 12% of patients had complaints of joint pain.
- Laboratory investigations showed anaemia in 25% and raised ESR in 57% of the patients.
- ANA was found in 29% of the patients. ANA positivity was more common in disseminated DLE(42%) than in localized DLE (26%).
- Anti-double stranded DNA was negative in all the patients.

- Rheumatoid factor was positive in 33% of the patients. C reactive proteins was positive in 33% of the patients.

- Histopathological study showed compatibility with the features of DLE in all cases, except one. One case showed features of lichen planus. Another case showed features of Verrucous DLE.

- Direct immunofluorescence study showed moderately strong homogenous IgG, IgM, IgA and C3 at the basement membrane zone band in the lesional skin and negative findings in the uninvolved skin, features suggestive of Discoid Lupus Erythematosus in most of the cases. One case showed discontinuous ragged fibrinogen band in the basement membrane zone and IgG, IgM and C3 colloid bodies in the lesional skin. These features were suggestive of Lichen planus.

- Neoplastic change was noted in 2% of the cases.

Figure 1

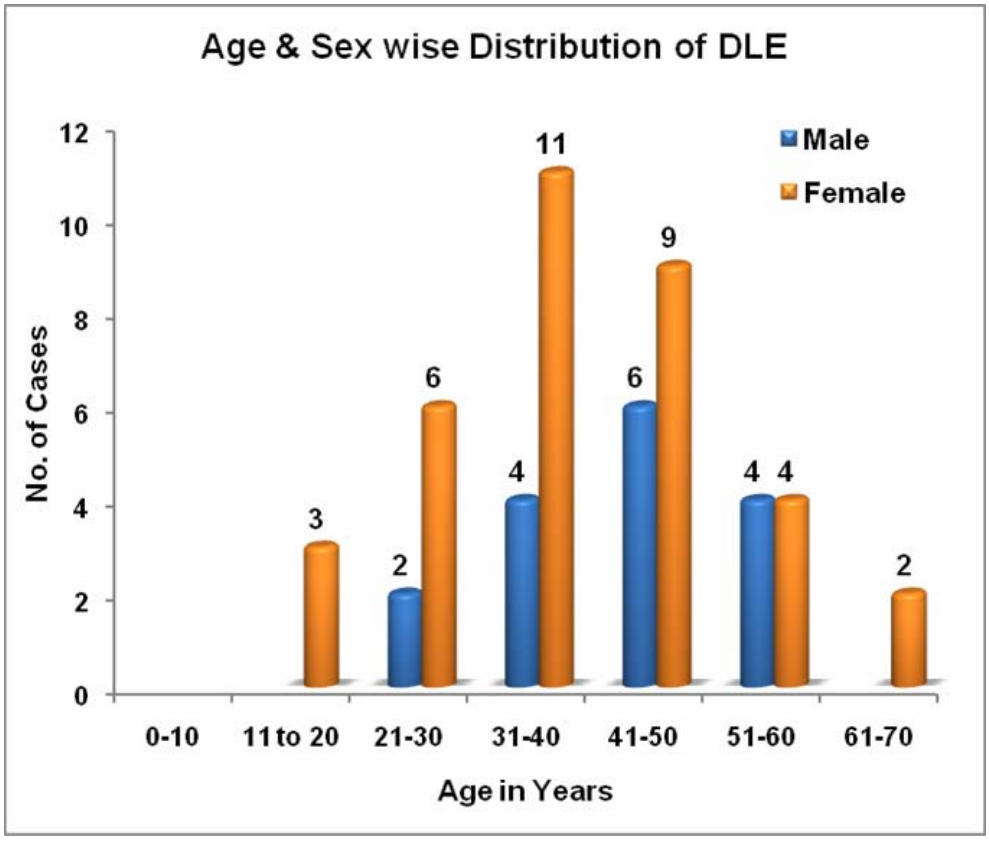

Age & Sex wise Distribution of DLE
Male
Female
No. of Cases
12
10
8
6
4
2
0
3
2
6
4
11
6
9
4
4
2
0-10
11 to 20
21-30
31-40
41-50
51-60
61-70
Age in Years

Figure 2

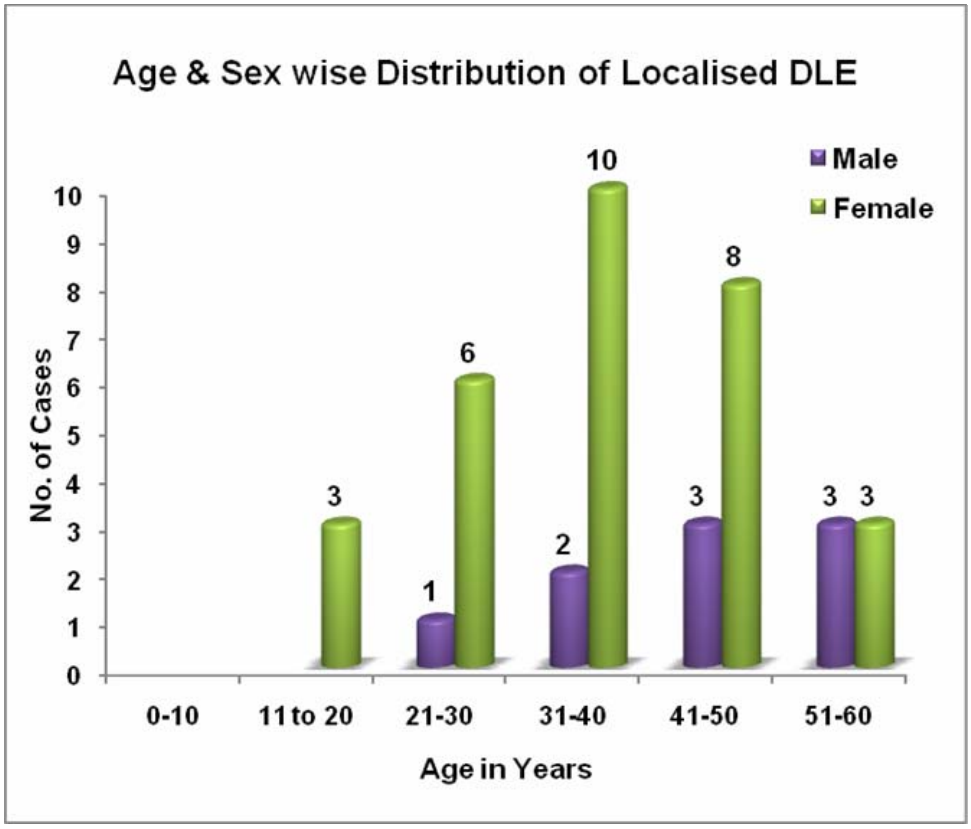

Age & Sex wise Distribution of Localised DLE
Male
Female
No. of Cases
0
1
2
3
4
5
6
7
8
9
10
3
1
6
2
10
3
8
3
3
0-10
11 to 20
21-30
31-40
41-50
51-60
Age in Years

Figure 3

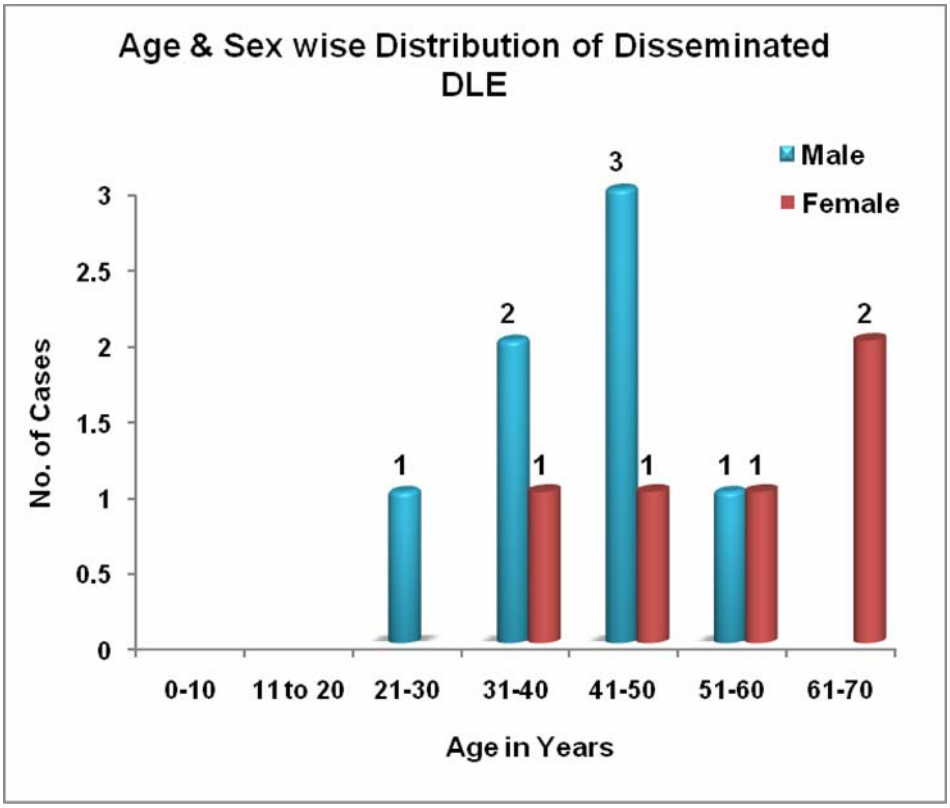

Age & Sex wise Distribution of Disseminated DLE
Male
Female
No. of Cases
0
0.5
1
1.5
2
2.5
3
1
2
1
3
1
1
1
2
0-10
11 to 20
21-30
31-40
41-50
51-60
61-70
Age in Years

Figure 4

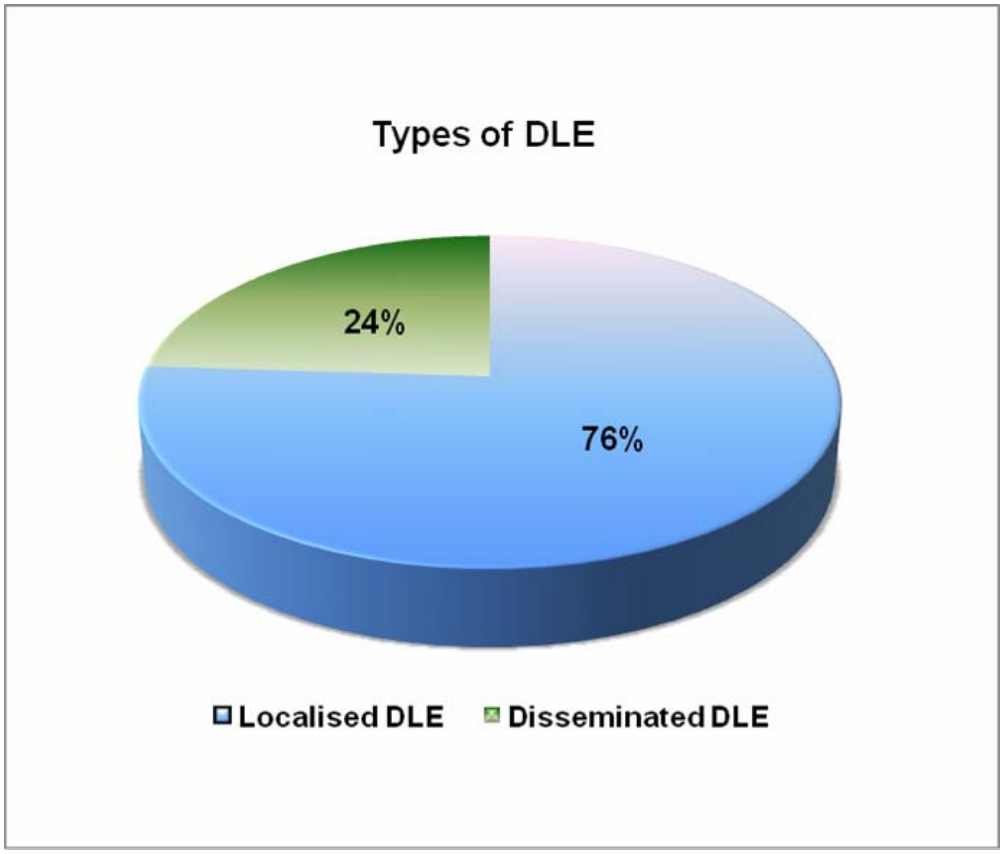


Figure 5

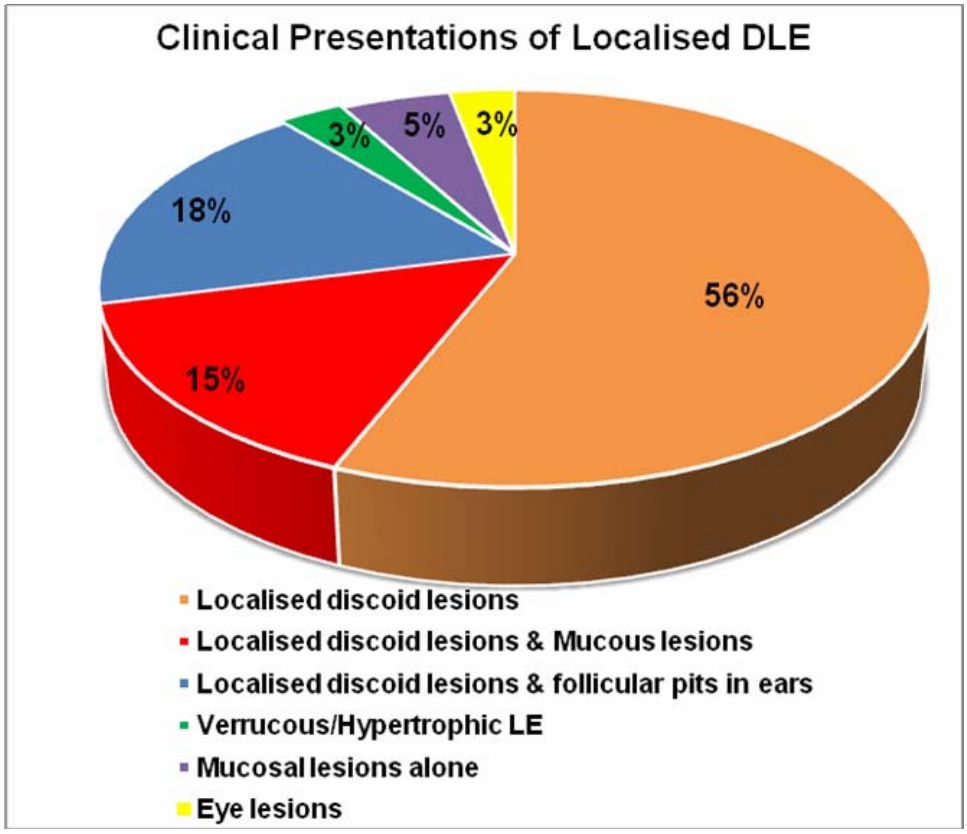

Clinical Presentations of Localised DLE
56%
15%
18%
3%
5%
3%
Localised discoid lesions
Localised discoid lesions & Mucous lesions
Localised discoid lesions & follicular pits in ears
Verrucous/Hypertrophic LE
Mucosal lesions alone
Eye lesions

Figure 6

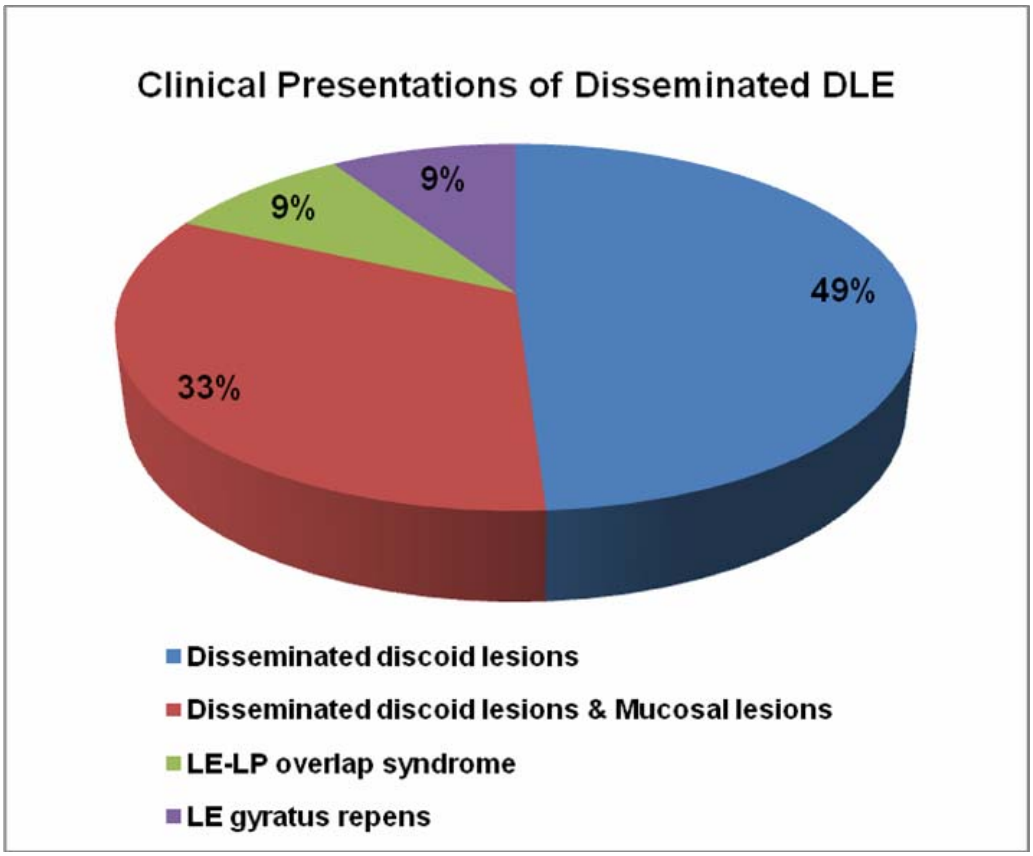
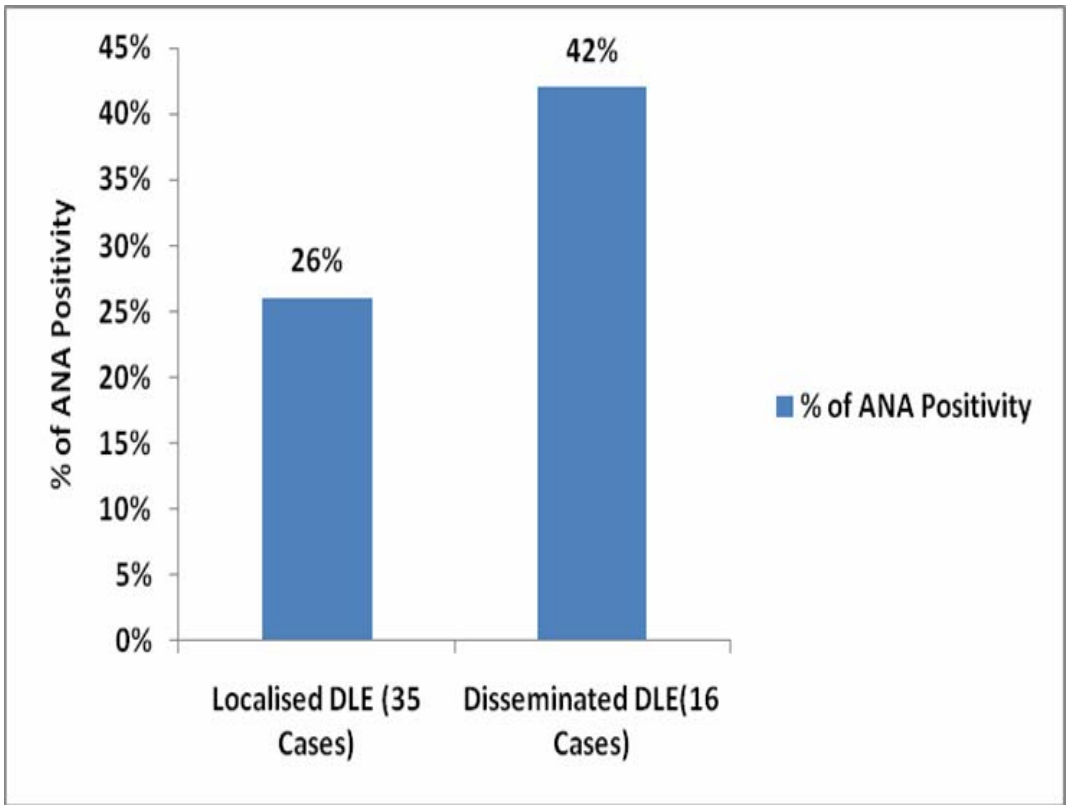

Clinical Presentations of Disseminated DLE
49%
33%
9%
9%
Disseminated discoid lesions
Disseminated discoid lesions & Mucosal lesions
LE-LP overlap syndrome
LE gyratus repens

Figure 7


45%
40%
35%
30%
25%
20%
15%
10%
5%
0%
% of ANA Positivity
26%
42%
Localised DLE (35 Cases)
Disseminated DLE(16 Cases)
% of ANA Positivity

LOCALISED DLE
ERYTHEMATOUS SCALY ATROPHIC PLAQUES

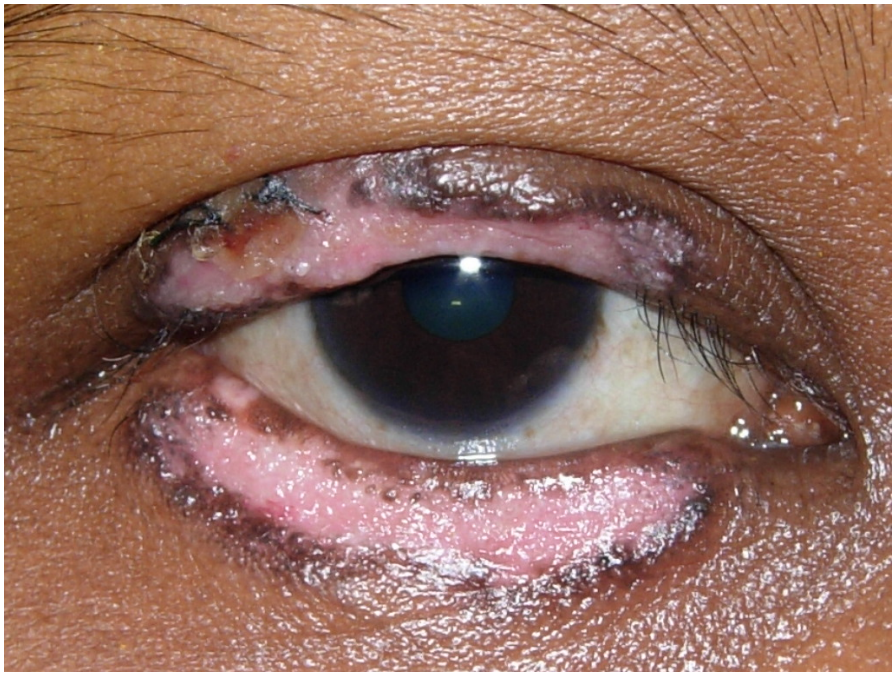
LOCALISED DLE WITH SCARRING ALOPECIA


29.01.2008 09:52

DISSEMINATED DLE

DISSEMINATED DLE

DLE – LESIONS OVER THE EYELID



FOLLICULAR PITS IN THE EAR

MUCOSAL DLE – LESION OVER THE LIPS

MUCOSAL INVOLVEMENT – HYPERPIGMENTED PATCHES OVER THE BUCCAL MUCOSA

VERRUCOUS / HYPERTROPHIC DLE

LE / LP OVERLAP SYNDROME WITH SQUAMOUS CELL CARCINOMA

SQUAMOUS CELL CARCINOMA IN A CASE OF LE / LP OVERLAP SYNDROME

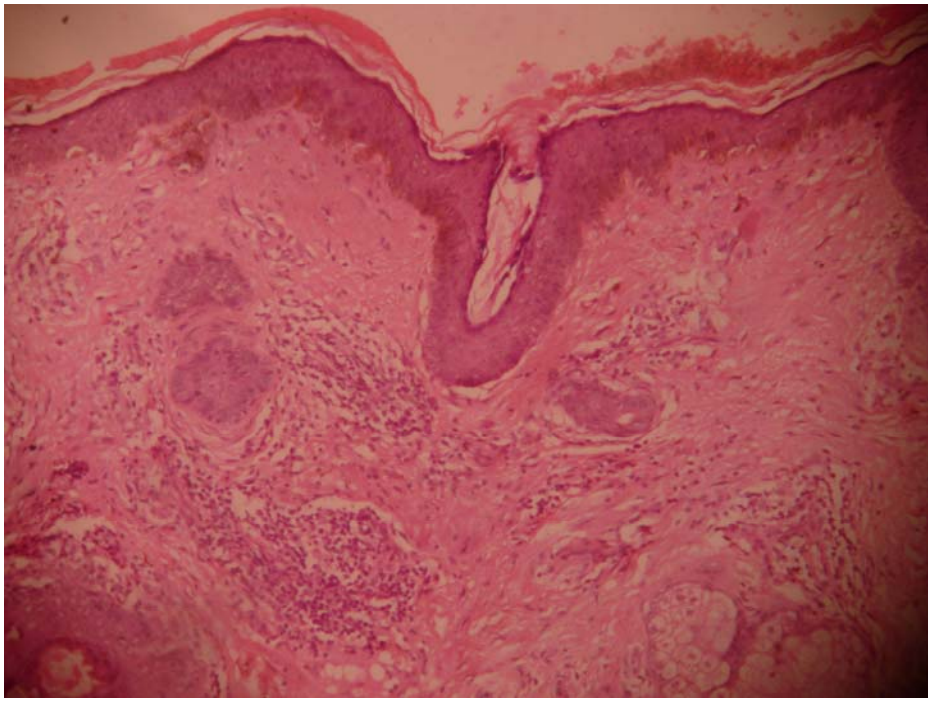
LE GYRATUS REPENS

LE GYRATUS REPENS

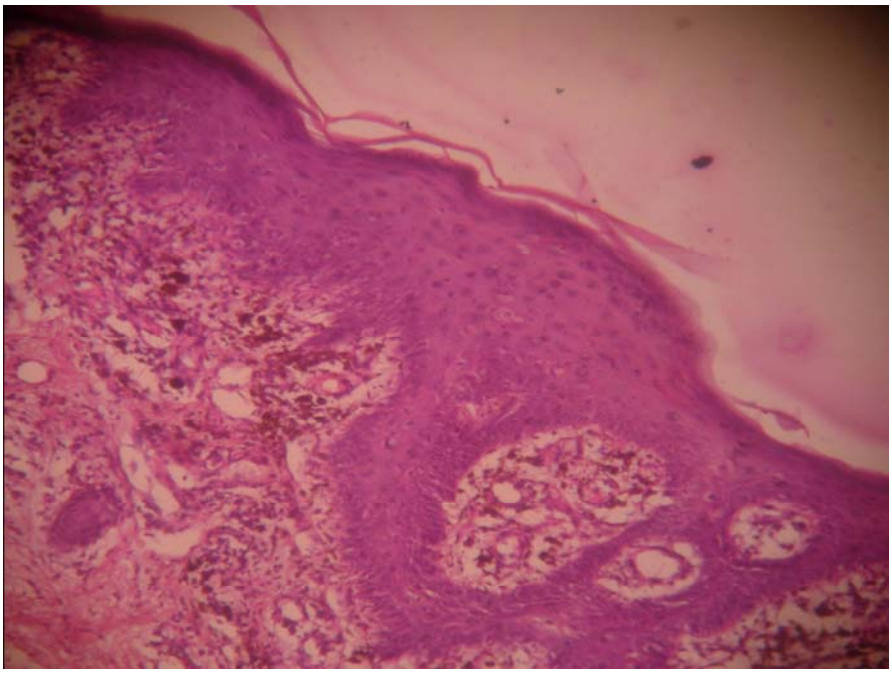
DLE WITH PRURIGO NODULARIS

DLE ASSOCIATED WITH POLYMORPHOUS LIGHT ERUPTION

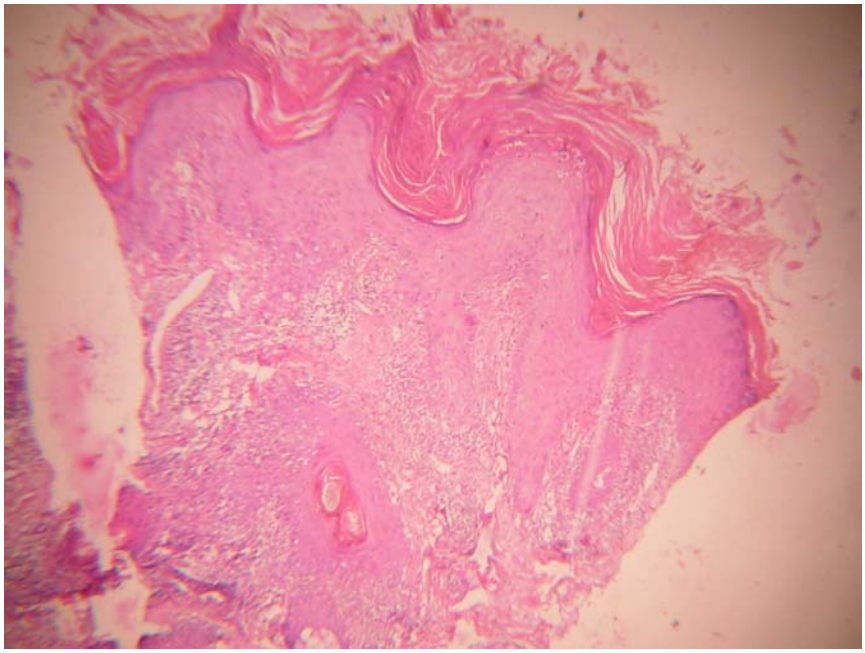
HISTOPATHOLOGY OF DLE – SHOWING FOLLICULAR PLUGGING BASAL CELL DEGENERATION & PATCHY INFLAMMATORY INFILTRATE IN DERMIS



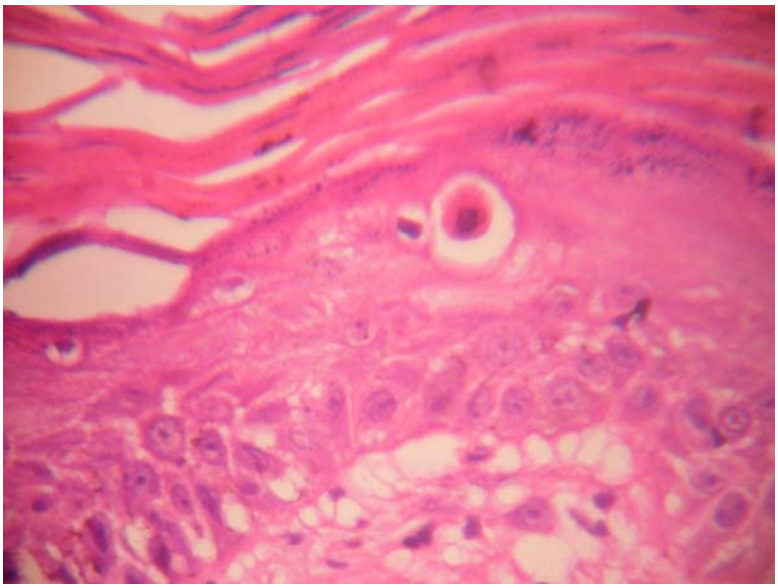
BASAL CELL DEGENERATION WITH PIGMENT INCONTINENCE



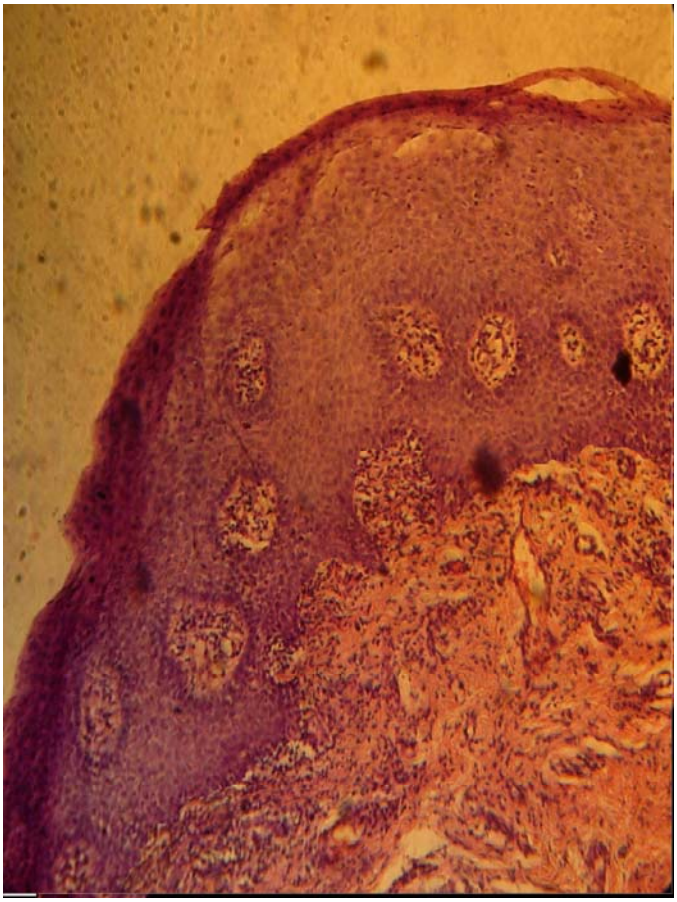
HISTOPATHOLOGY OF HYPERTROPHIC / VERRUCOUS DLE : SHOWING HYPERKERATOSUS, PAPILLOMATOSIS, BASAL CELL DEGENERATION & PERIAPPENDAGEAL INFLAMMATORY INFILTRATE



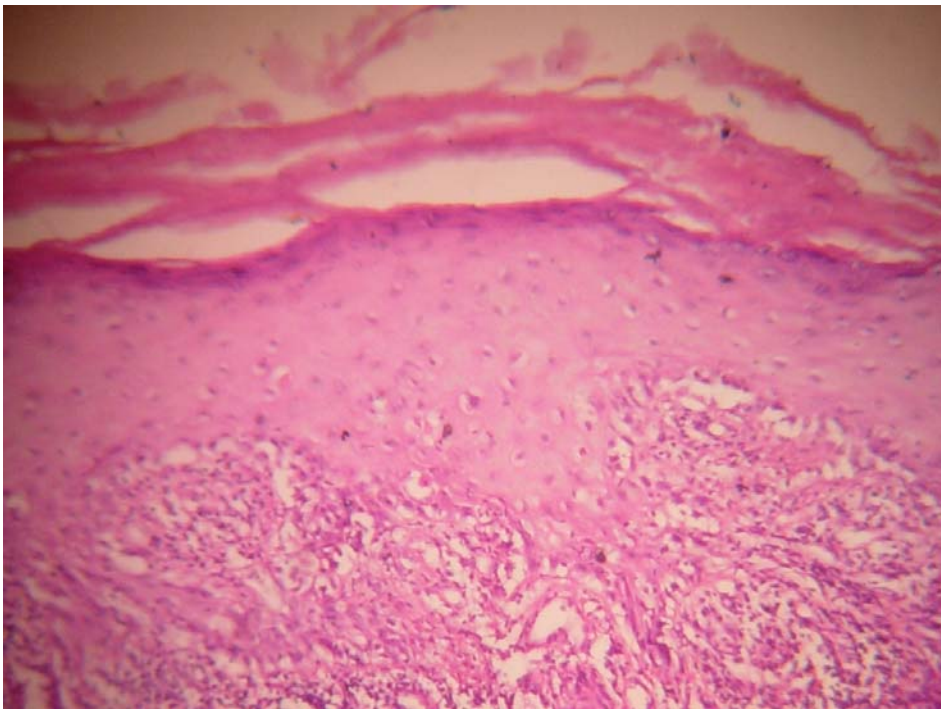
COLLOID BODY – HIGH POWER VIEW



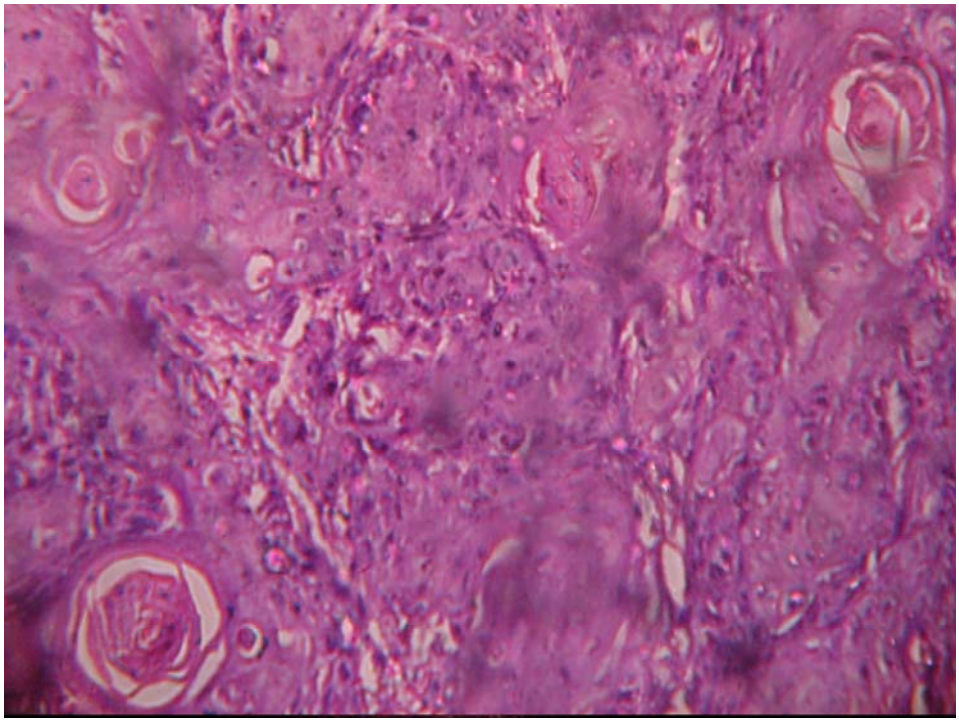
MUCOSAL DLE – BASAL CELL DEGENERATION WITH PATCHY INFLAMMATORY INFILTRATE IN DERMIS



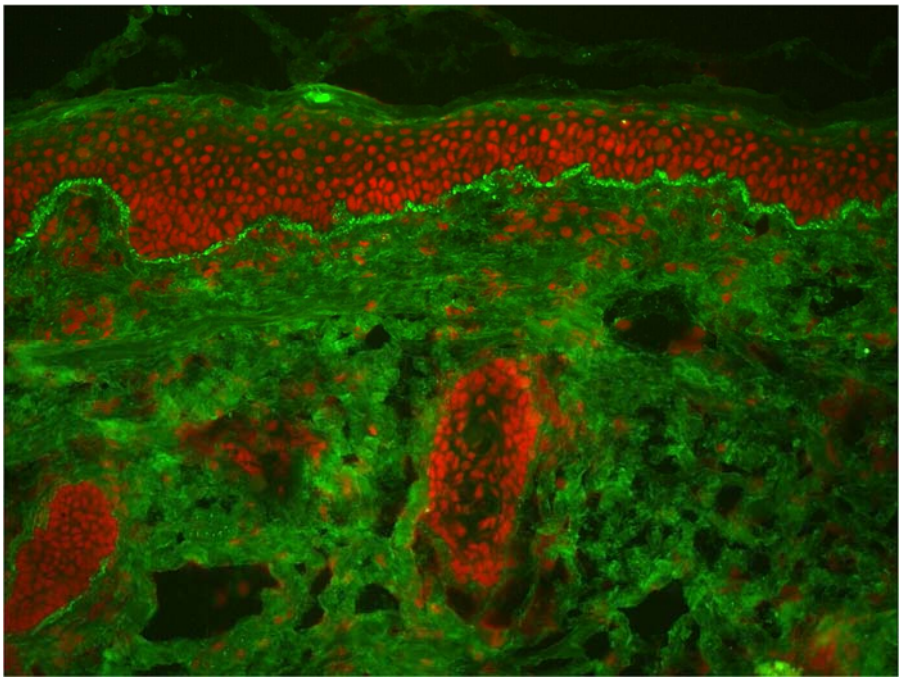
LE / LP OVERLAP SYNDROME : HPE SHOWING FEATURES OF LICHEN PLANUS



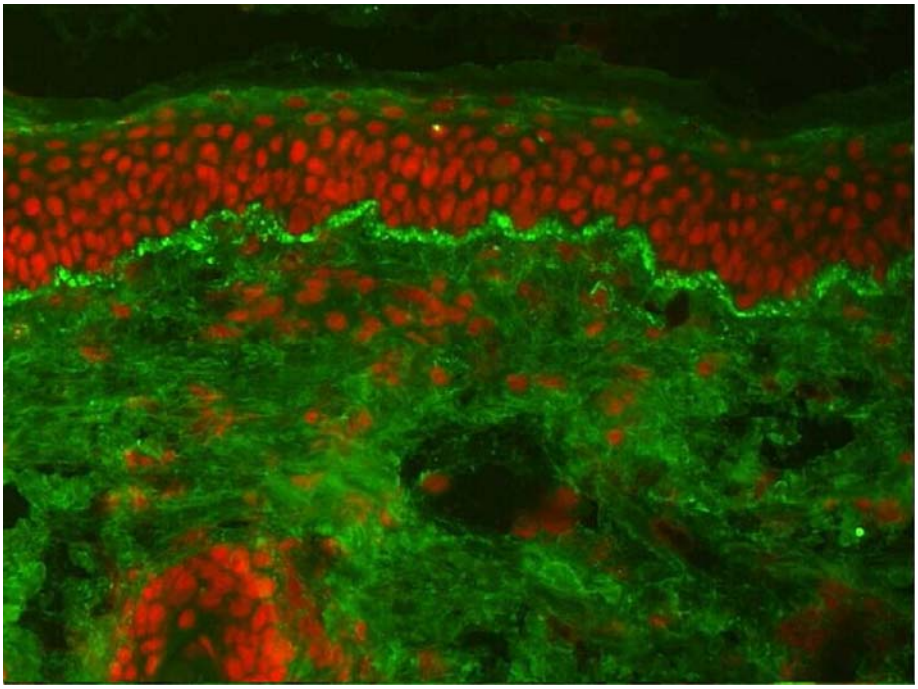
SQUAMOUS CELL CARCINOMA- HPE SHOWING HORN PEARLS



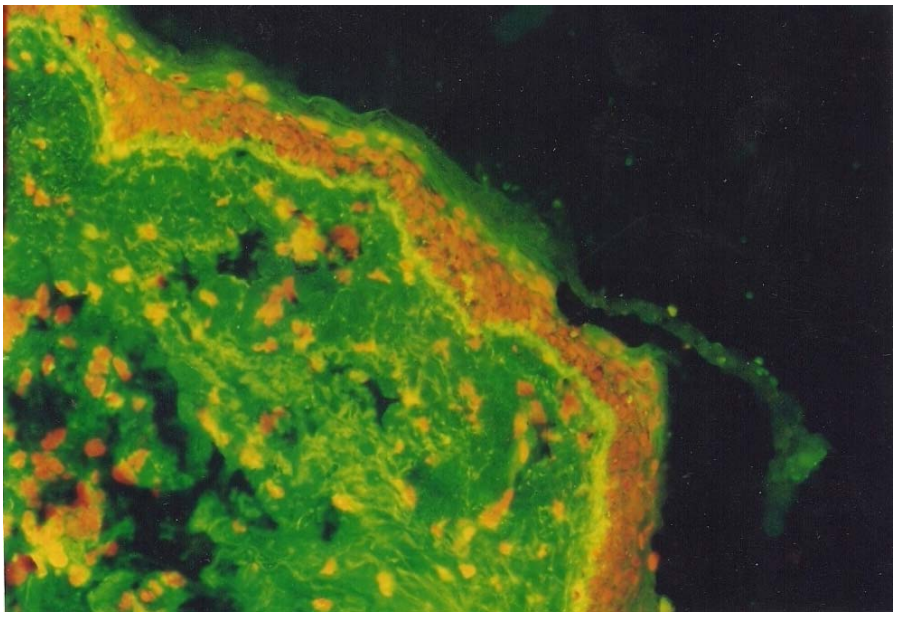
DIRECT IMMUNOFLUORESCENCE – SHOWING MODERATELY STRONG HOMOGENOUS BAND OF IgG AT THE BASEMENT MEMBRANE ZONE IN THE LESIONAL SKIN



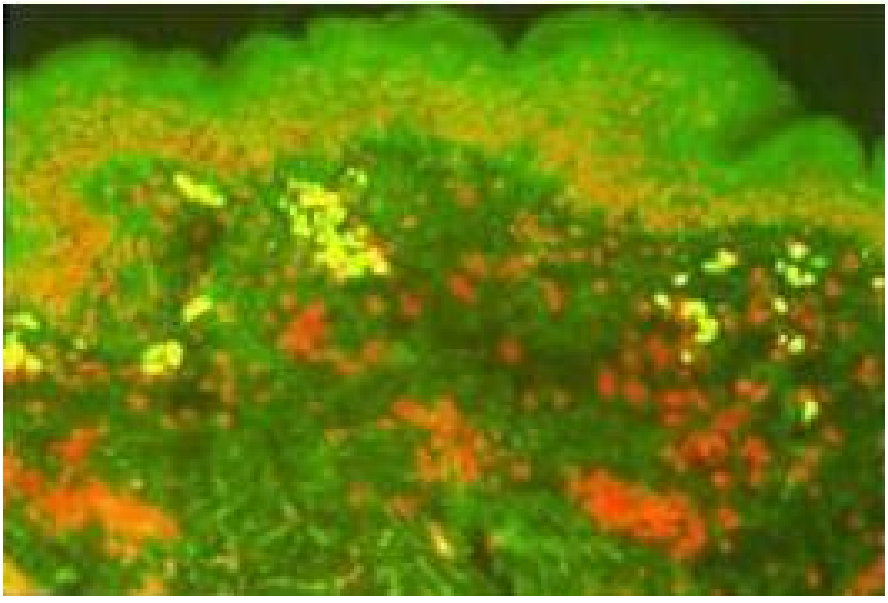
DIF-SHOWING MODERATELY STRONG HOMOGENOUS BAND OF C3 AT THE BASEMENT MEMBRANE ZONE IN THE LESIONAL SKIN



DIF-SHOWING HOMOGENOUS BAND OF IgM AT THE BASEMENT MEMBRANE ZONE IN THE LESIONAL SKIN



DIF – IgG COLLOID BODIES IN THE LESIONAL SKIN



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PROFORMA

Case no. **O.P No.**

Complaints :

Duration : **Onset :**

SITE

Head : Scalp , Face , Ears , Eyes , Lips , Oral mucosa, nasal mucosa

Trunk : Chest , Back , Abdomen, Genitalia

Limbs : Upper limbs / Lower limbs , Palms , Soles

ASSOCIATED COMPLAINTS

Photosensitivity: Itching: Pain: Scaling: Raynaud's Phenomenon: Joint Pain:

PRECIPITATING FACTORS

Sunlight: Drugs: Dapsone, Isoniazid, Griseofulvin, Penicillamine

Trauma: Radiation exposure: Mental stress:

Exposure to cold: Pregnancy: Premenstrual deterioration:

HISTORY RELATED TO SYSTEMIC INVOLVEMENT:

Constitutional symptoms, Arthralgia, Chest pain, Palpitation, Dyspnea, Dysphagia, Vomiting, Abdominal pain, Epilepsy, Psychosis, Oliguria, Pedal edema.

PAST HISTORY:

PERSONAL HISTORY: Smoking, Alcohol

MENSTRUAL HISTORY (In Female): Any Premenstrual exacerbation

OCCUPATIONAL HISTORY:

TREATMENT HISTORY:

GENERAL EXAMINATION:

Anaemia: Jaundice: Cyanosis: Pedal edema: Lymphadenopathy: Clubbing:

Vital signs: Pulse rate: BP: TEMP: RR:

SYSTEMIC EXAMINATION:

C.V.S: R.S: C.N.S: PER/ABD:

DERMATOLOGICAL EXAMINATION:

MORPHOLOGY: Plaque/Patch: Erythema: Depigmentation: Atrophy: Scaling: Hyperpigmentation: Scarring:

Carpet Tacks Sign:

SITE:

LOCALISED:

Scalp: Face: Ears: Neck:

Retroauricular: `Oral mucosa: Nasal mucosa: Lips: Eyes:

DISSEMINATED:

Chest: Back: Arms: Forearms: Dorsum of hand:

Palms: Thighs: Legs: Dorsum of Foot: Soles:

Genitalia:

Nails:

Hair: Alopecia

INVESTIGATION:

Urine: albumin, sugar, deposits.

Hb: Total count: Differential count: ESR:

Platelet count:

Renal Function Test: Bd Sugar, Urea, Serum Creatinine

Liver Function Test: Serum Bilurubin,Total Proteins, SGOT, SGPT, Alkaline phosphatase.

ANA Titre:

Rheumatoid factor: C-reactive protein:

Histopathological Examination:

Direct immunofluorescence:

Rheumatology opinion:

KEY TO MASTER CHART

M	- Male
F	- Female
Ppt	- Precipitating factor
Aggr	- Aggravating factor
Jt	- Joint
Clin	- Clinical
Asst	- Associated
Hb	- Haemoglobin
TC	- Total count
ESR	- Erythrocyte sedimentation rate
RFT	- Renal function test
LFT	- Liver function test
RF	- Rheumatoid factor
CRP	- C reactive protein
ANA	- Antinuclear antibody

ds-DNA	- Double stranded DNA
S	- Smoking
A	- Alcohol
Yrs	- Years
Agri	- Agriculture
H.W	- Housewife
W.M	- Watchman
S.K	- Shopkeeper
Stud	- Student
Driv	- Driver